The Ear: Audition and Balance

Introduction

This lesson focuses first on the normal vestibulocochlear apparatus and then on disorders of audition and balance. Throughout the lesson, you will see that both hearing and balance function through the use of **hair cells**. Each system uses them a little differently, but they all rely on the same underlying mechanism—the hairs, called **stereocilia**.

Audition involves the **translation of sound waves into axonal action potentials**. Sound waves are directed towards the middle ear by the external ear, then translated from a sound wave to a vibration of the middle ear ossicles. The vibration is delivered to the cochlea (the portion of the vestibulocochlear apparatus that is involved in hearing), which translates that vibration into the electrical signal sent to the brain along axons that make up the cochlear fascicles of the cochlear nerve.

Balance is managed by the vestibular portion of the vestibulocochlear apparatus and translates movement in space to an electrical impulse, carried in the vestibular portion of the vestibulocochlear nerve. This part of the organ relies on gravity to assess a relative position change. Each of the rotation movements (x, y, z planes) and directional movements (anterior, posterior, lateral) is coded by a continuum of hair cells.

The vestibule and cochlea are related anatomically, but not functionally. They share the special fluid called endolymph and are within the same protective coating, but the two parts don't have a physiological interaction. The cochlear neurons and their axons don't interact with the vestibular neurons and axons. This lesson's organization—audition, start to finish, and then balance, start to finish—keeps the two functions mentally separate in your learning, helping to solidify in your memory that all the structures, features, and pathology of one are separate from those of the other. But because they both rely on hair cells, we kept them in the same lesson. (We can also now give you challenge questions that can trip you up if you form loose associations instead of strong memories.)

The Anatomy of Audition and Balance

We're going to go hard and fast on the anatomy. You need to see the structures and understand their relationships, but most of what we're going to talk about is the inner ear, not the outer or middle ear. Use this illustration to follow along.

The pinna (the thing you call your ear when you look in the mirror) is lined with skin and made of fibroelastic **cartilage**, much like the exterior nose. You can bend your ear, and it will snap back to its original shape. The pinna, also known as the **auricle**, is designed to capture sound waves and project them into the external auditory canal. The **external auditory canal** extends from the auricle to the tympanic membrane. It is only 2.5 cm long and is also lined with skin. The opening of the canal is made of cartilage, the same as the pinna, but quickly becomes encased in the temporal bone. The outer ear is ectoderm-derived and lined with skin—hair-bearing, keratinized, stratified squamous epithelium. That epithelium has **ceruminous glands** that produce cerumen, aka earwax. Just as mucus produced by submucosal glands protects the respiratory epithelium, the hairs and cerumen prevent organisms from reaching the medial boundary of the outer ear, the tympanic membrane.

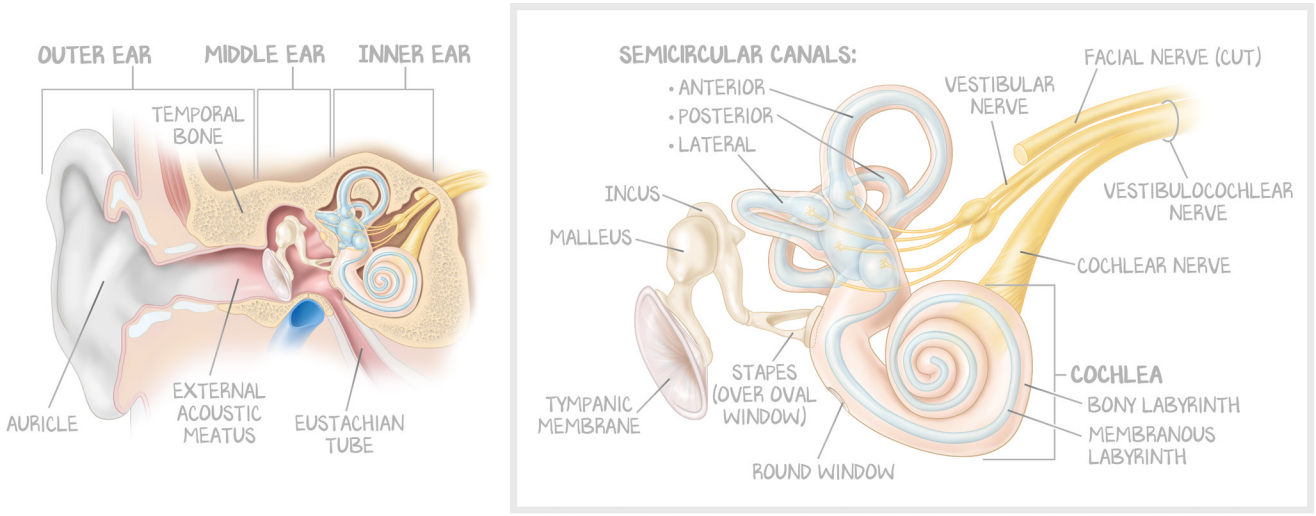

Figure 4.1: Anatomy of Audition and Balance
The ear is divided into the outer ear (the pinna to the tympanic membrane), middle ear (the gut-tube derived, respiratory epithelium-lined cavity in which the bones of audition are located), and inner ear (where the vestibulocochlear apparatus is located). Although they form one continuous structure, the vestibule is responsible for spatial perception and rotational movement, whereas the cochlea is responsible for the perception of sound. They both utilize perilymph and endolymph, and each structure transmits electrical impulses along cranial nerve VIII, the vestibulocochlear nerve. The malleus transmits sound vibration from the tympanic membrane onto the incus, the incus onto the stapes, and the stapes onto the oval window.

The **tympanic membrane** is a connective-tissue membrane that separates the **outer ear** (auricle and external auditory canal) from the middle ear. The **middle ear** is lined with respiratory epithelium—**simple columnar epithelium** with both ciliated columnar cells and goblet cells. The middle ear technically includes the **tympanic cavity**, where the ossicles of audition (the bones that make sound perception possible) are located, and the **eustachian tube**, which connects the oropharynx to the tympanic cavity. In the ENT section of Pulmonary, we talked about the eustachian tube as an appendage of the oropharynx, likening it to the other gut tube appendages—the trachea and nasal cavity—to reinforce the concept that similar pathogens would infect similar epithelia. There, we didn't mention the tympanic membrane, saving it for the discussion here.

The **tympanic cavity** is a large, open structure, into which the eustachian tube opens. The eustachian tube is designed to open upon yawning or swallowing. Because the tympanic membrane makes a complete seal between the outer ear and middle ear, the only connection to the atmosphere is through the eustachian tube and oropharynx. The eustachian tube **equalizes the pressure in the middle ear** to that of the surrounding atmosphere, enabling the bones of audition (those bones' functions are described in the next section) to work more effectively. The tympanic cavity is bounded on six sides: anterior, posterior, medial, lateral, superior, and inferior. Although lined with epithelium and separated from the bone itself, the tympanic cavity develops within the temporal bone. The **posterior wall** is the **mastoid process** of the temporal bone. The **anterior** is the temporal bone, through which the eustachian tube enters the cavity. The superior and inferior aspects are both thin strips of the temporal bone that separate the tympanic cavity from the jugular vein (inferior) and carotid artery (superior). The **lateral** boundary is the **tympanic membrane**. The **medial** boundary is the bony wall of the temporal bone that houses the structures of the inner ear (also discussed in the next section). Within that bone are two openings that connect the middle ear to the inner ear, both related to sound—the **oval window** and the **round window**. This is going to be where vibrations "enter" (oval window) and "exit" the cochlea (round window). The tympanic cavity houses the three small bones of audition, the ossicles.

The **inner ear** is neuroectoderm-derived neural tissue and is connected to the brain by the **vestibulocochlear nerve**. The "inner ear" is better described as "the eye of hearing" than "the inner ear," but because there are three distinct cavities—outer ear with skin, middle ear with respiratory epithelium, then the organs of hearing and balance in the bone—the convention is outer, middle, inner. The ossicles connect to the **cochlea**, the swirling snail-shell-shaped portion of the inner ear that translates the **mechanical** signals from the middle ear into **electrical** signals (action potentials), thereby translating conducted vibration into perceivable sound. The **vestibular apparatus** is the organ of balance and is separated into the **semicircular canals** and the **vestibule**. The vestibular portion is discussed after the cochlea, later in this lesson.

Physiology of Audition

The tympanic cavity houses the **ossicles** of the middle ear—three small bones that conduct and amplify sound, named the malleus, incus, and stapes. The **malleus** (hammer) is connected to the tympanic membrane, articulates with the incus, and translates the sound waves vibrating the tympanic membrane into an osseous signal. The **tympanic membrane** receives the sound waves, vibrates, and the vibration moves the hammer, which strikes the anvil. The **incus** (anvil) is the largest of the three ossicles and connects the malleus to the stapes. The **stapes** (stirrup) is a long slender bone, the footplate of which (which is in the shape of a stirrup of a horse saddle) fits into the oval window, where it acts as a piston on the cochlear fluid. The malleus sends the signal from the tympanic membrane, the stapes sends the signal to the inner ear, and the incus is the middleman, used as an extra layer through which sound can be modified. In addition to the incus, there are two muscles, the **tensor tympani** (which inserts on the tympanic membrane) and the **stapedius** (which inserts on the stapes). Contraction of these muscles makes the chain of ossicles more rigid, limiting conduction through the system by preventing vibration of the tympanic membrane, protecting the inner ear from loud noise. See Figure 4.1 for details.

The vestibulocochlear unit is made of a **bony labyrinth** (the word labyrinth describes the many twists in its shape, and is the word that means "*hearing and balance thingamajigger*") and a **membranous labyrinth**. The bony labyrinth is the outer case made from bone, which protects the inner membranous labyrinth. Within the cochlea, perilymph circulates through the outer chambers, and endolymph flows through the center channels. In the vestibular unit, the same endolymph flows through the semicircular canals and around the saccule and utricle. We turn your attention to only the organs of audition—the cochlea and the organ of Corti.

The **cochlea** is responsible for translating a mechanical signal—vibration—into an electrical one—action potentials. The arrangement of the cochlea is immensely complicated. We didn't have enough room to fit in all the details, so we left a lot out. The cochlea is best understood if it is unraveled from its snail-shell-like appearance and splayed out flat. The stapes translates vibration into a canal of perilymph through the oval window. That canal passes from the base to the apex, takes a turn around the apex, and returns to the base, arriving at the round window. This perilymph tunnel surrounds the **cochlear duct**, the **endolymph-filled** chamber that houses the **organ of Corti**, which is responsible for translating vibration into action potentials.

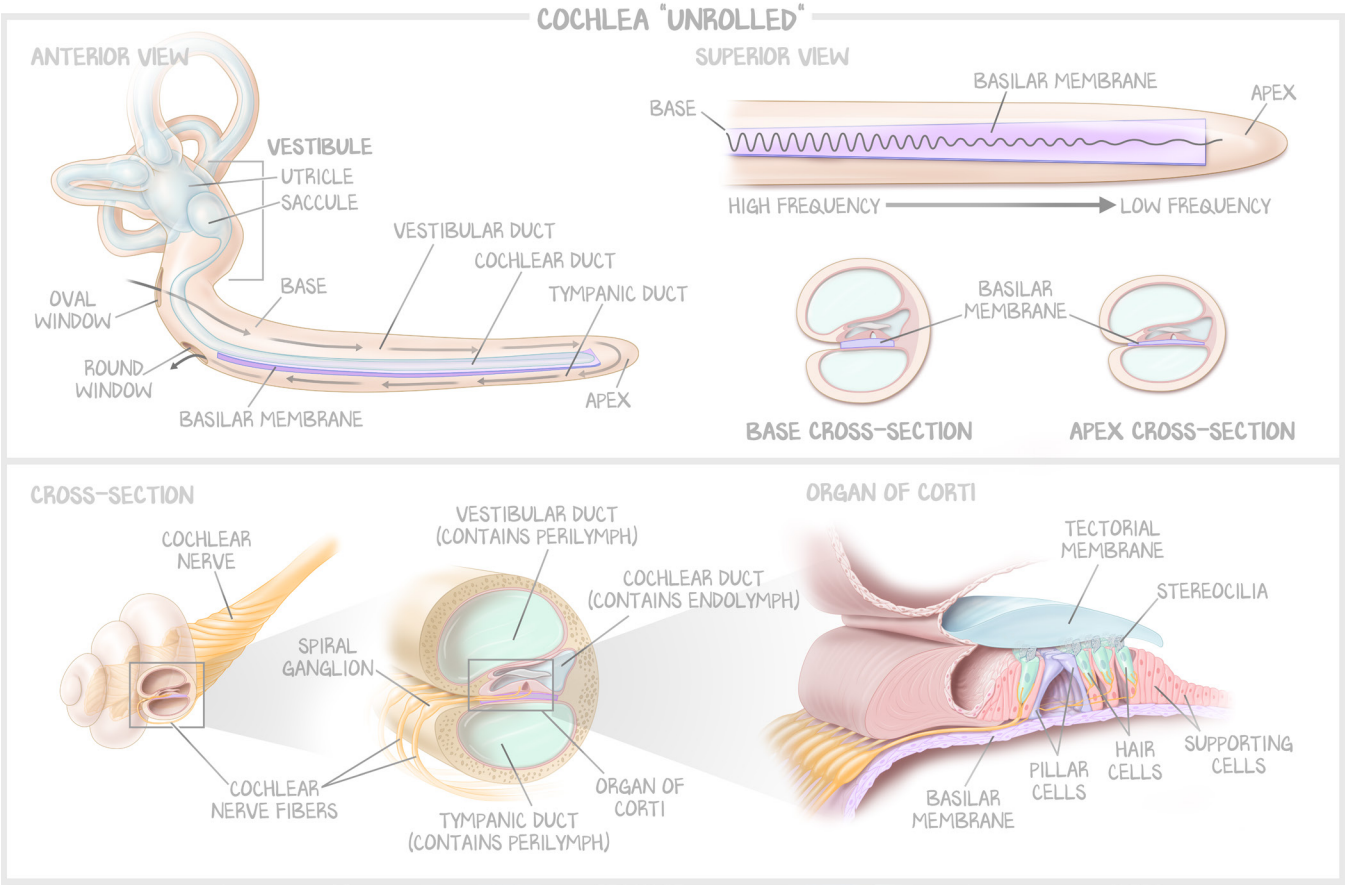

Figure 4.2: Anatomy and Histology of the Cochlea
We have illustrated the cochlea as "unrolled" to demonstrate the change in the basilar membrane. At the start of the cochlea, the basilar membrane is tall and skinny. At the end, it is flat and wide. In between, there is a gradation from one to the other. This enables the membrane to vibrate more or less vigorously in a continuous manner across the spectrum of human audition. The hair cells have stereocilia in the tectorial membrane and remain fixed there. The basilar membrane vibrates, shifting the cell bodies from below.

The **basilar membrane** of the organ of Corti is what the epithelium, hair cells, and axons that transmit action potentials are anchored upon. The **tectorial membrane** is fixed and to what the hair cells' **stereocilia attach.** These things matter, but let's focus on the basilar membrane first. The basilar membrane will vibrate in response to the stapes' vibration of perilymph. The **basilar membrane** starts **narrow and thick**, tuned for high-frequency sound, at the base of the cochlea, where vibrations both enter and exit perilymph. The basilar membrane of the organ at the **apex** is **wide and thin**, best tuned for low frequencies. There is a gradual shift from the base to the apex, meaning that the spectrum of sounds a human can hear is represented spatially along the cochlea. When the stapes vibrates the perilymph, the basilar membrane will vibrate, and vibrate the most at the location along its length that most closely matches the stapes' frequency. The basilar membrane vibrates, but the tectorial membrane doesn't. The epithelium is drastically simplified here for brevity. The basilar membrane is the foundation of the epithelium. The epithelium consists of a **bipolar neuron** layer (**pillar** cells) and a **hair cell** layer. The hair cells are attached to the tectorial membrane by **stereocilia**. These stereocilia have decreasing lengths, tallest at the most lateral apex of the cell, shortest at the most medial. Every hair cell has this arrangement. When the basilar membrane vibrates, because the basilar membrane moves but the tectorial membrane doesn't, there is a shearing force felt by the stereocilia, translated through the hair cell to the pillar cell. The pillar cells' axons form the cochlear nerve, which exits the organ with the vestibular nerve as the vestibulocochlear nerve.

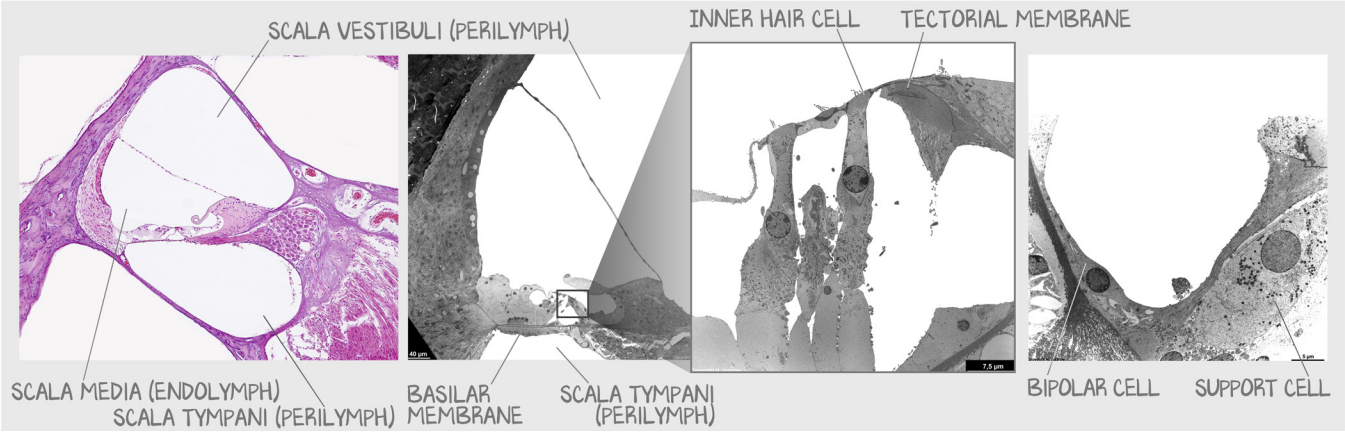

Figure 4.3: Anatomy and Histology of the Cochlea
(a) Light microscopy showing the organ of Corti and the different scala, as well as the nerve exiting to the right from the center. (b) Electron microscopy of the same thing, with limited views of the scala vestibuli and scala tympani but a focus on the basilar membrane. (c) Higher magnification of the electron microscopy in panel b demonstrating the connection of the hair cells (only the inner hair cell is labeled) to the tectorial membrane. (d) Electron microscopy of the bipolar cell's projection off to the right (the hair cell would be up and to the left).

The signal is translated into an electrical impulse through **potassium channels**. In every other excitable cell that we have ever taught you, a potassium channel has been responsible for hyperpolarizing a cell. **Endolymph is special** because of its **extraordinarily high potassium** concentration. When the hair cells are deflected, held in place by the tectorial membrane but vibrated by the basilar membrane, the hair cells are displaced, revealing potassium channels. **Potassium rushes into the cell**, leading to a **depolarizing signal**. Depolarization opens voltage-gated calcium channels on the presynaptic (hair cell) side of a synapse, induces vesicle fusion, and releases glutamate onto the pillar cell, which depolarizes and sends its signal on axons that project to the thalamus and parietal lobe.

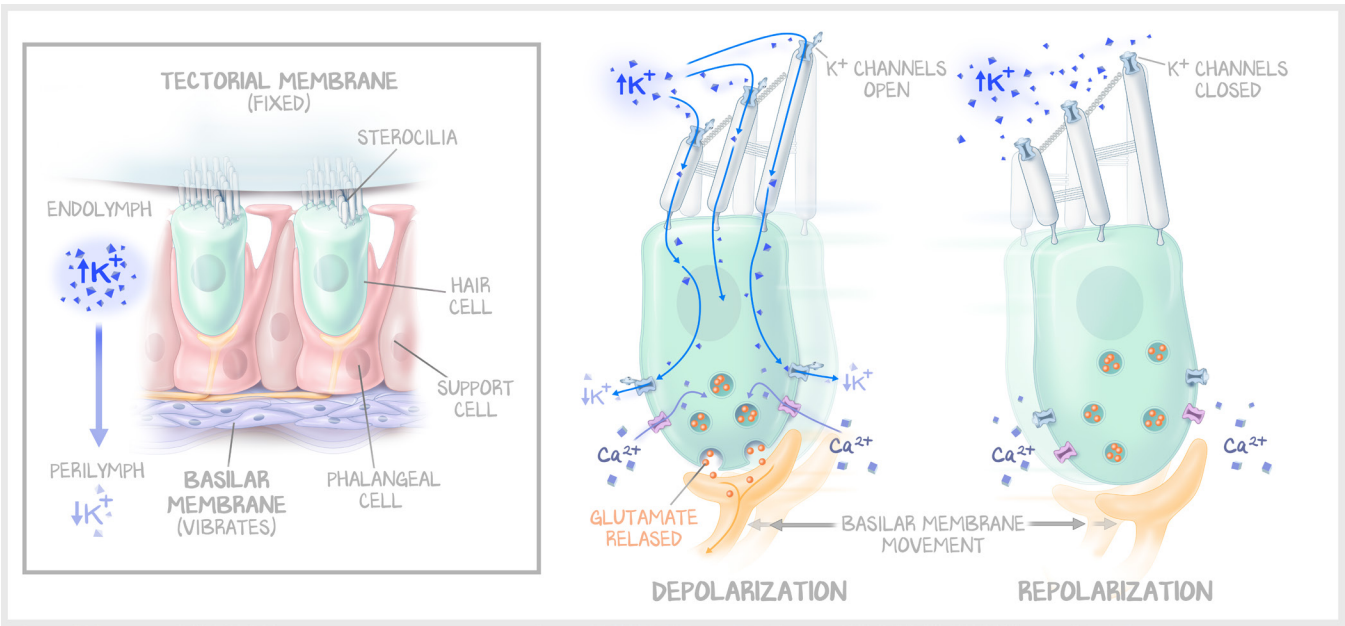

Figure 4.4: Hair Cells and Potassium
The tectorial membrane vibrates. The hair cells stay fixed in the tectorial membrane. The movement of the stereocilia creates either a more acute angle (closing the potassium channels) or a more obtuse angle (opening the potassium channels). Potassium channels lead to the movement of potassium from the endolymph into the cells, depolarizing them, which culminates in the release of glutamate onto the bipolar cells that connect the hair cells to the cochlear nerve neurons.

Hearing Problems

Problems with hearing come down to two types: conductive and sensorineural.

Conductive hearing loss is caused by any pathology that prevents the transmission of sound to the cochlea. From the cochlea to the perception of sound by the cortex is a problem of the brain, of neural tissue. But the sounds must first get from the atmosphere outside the person to the cochlea in order for sound to be interpreted. By definition, conductive hearing loss must be a pathology of the **outer or middle ear**.

Sensorineural hearing loss is impaired hearing due to either failure to translate (cochlea, basilar membrane) or failure to transmit (auditory nerve or auditory cortex) the electrical signal. The perception of sound, much like the perception of vision, requires a specific organ (cochlea) to translate vibrations into depolarizations. If at any point along the sensory tract, the tract of axons of neurons (sensorineural) or the nuclei those axons synapse onto are compromised, the signal never reaches its intended target, and so cannot be perceived.

Weber and Rinne Tests

The Weber test is used to determine laterality, and the Rinne test is used to determine conduction or sensorineural hearing loss. German physicians invented both of these physical exam maneuvers. The German Weber, in English, is approximated by "vaybuh" (vay as in vain, buh as in butter). The German Rinne, in English, is approximated by "rinnah" (as in "we'**RE IN A** heap of trouble!"). These are commonly appropriated into English as the "Webber" and "Rin" tests. Both pronunciations are accepted. But Rinne is definitely not French, so no "Rinays," unless you are talking about Renee Zellweger.

In conducting the physical exam, the **Weber** test is performed first. Striking a tuning fork against the examiner's hand induces vibration in the fork. The base of the fork is placed on top of the patient's head, at the midline. A **normal finding** is that the sound appears to be of equal volume and character on both sides. As the vibration weakens, the sensation of vibration will dwindle. A **normal finding** is that the perception of vibration ends at about the same time for both ears. **Abnormal findings** are a **disproportionate volume** between the two ears or **termination of perception in one ear before the other**. It does serve to **lateralize** the defect—whichever side is softer at the start or terminates first is the pathologic one that needs investigating. But it doesn't help separate conduction from sensorineural.

Perform the **Rinne** test on the side with **reduced sound**. Strike the tuning fork and place its base on the **mastoid bone** behind the ear. Ask the patient to report when the vibration can no longer be perceived. When they do, move the tuning fork to the side of the ear, held up and not touching the patient, so the tuning fork forms a U, pointed at the pinna. Ask the patient to report if any sound or vibration is perceived. And wait for that sound to be reported as absent (this last part ensures they really were hearing the tuning fork).

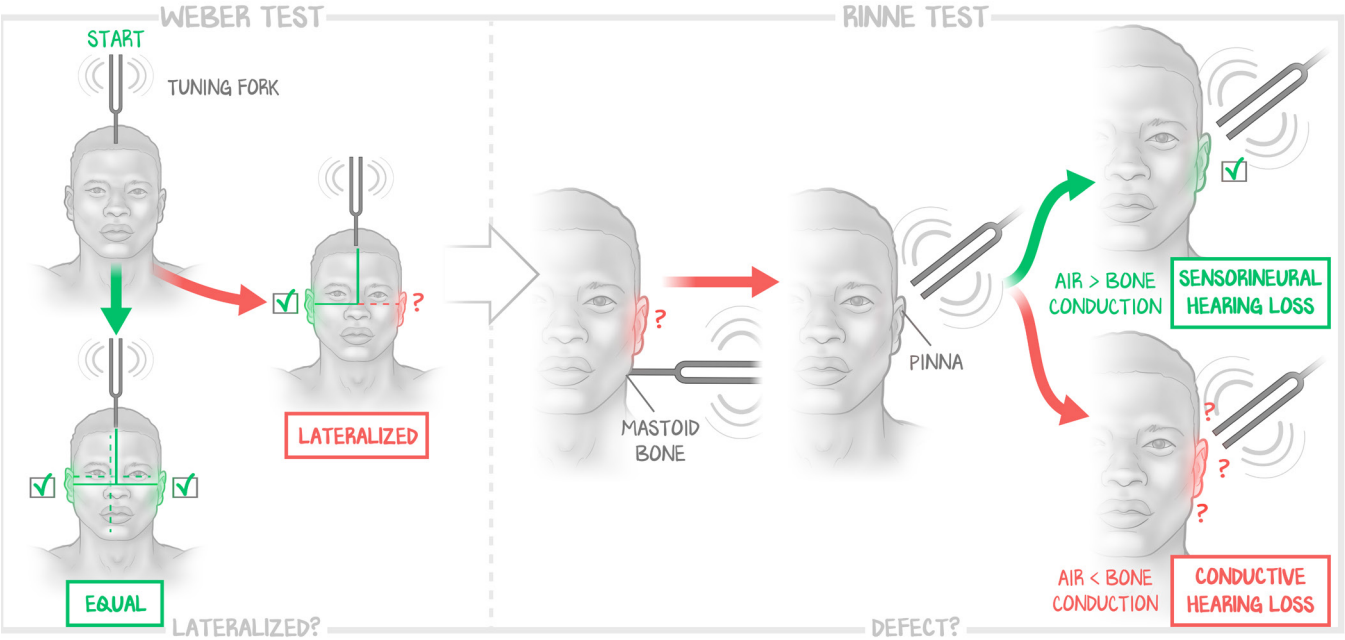

Figure 4.5: Weber and Rinne Tests
Start with the Weber test. Place the base of a vibrating tuning fork on the top of the head. If the vibration is interpreted as midline, stop. If one ear perceives less than the other, use the Rinne test to investigate the ear that detects less vibration. Place the base of a vibrating tuning fork on the mastoid process. When the patient reports they cannot hear any further vibrations, place the tuning fork, still vibrating, up to the auricle, the U shape directed down the auditory canal. If the sound is perceived, there is sound, and the defect is sensorineural. If no sound is perceived, the defect is conductive.

The tympanic membrane and ossicles are meant to translate vibrations to the cochlea. But that arrangement does more than just get the sound wave into a vibration—these structures also **amplify the signal**. The cochlea can sense vibration just through bone conduction. Therefore, if the patient lost perception at the mastoid, and then moving it to the ear regained the perception of sound, the amplifier is working, and it is not a conduction hearing loss. This is the case in normal ears and sensorineural hearing loss. But because the Weber test had already lateralized the reduced hearing in this ear, which rules out normal, the patient has **sensorineural hearing loss**. If the patient lost perception of the tuning fork from the mastoid and moving it to the ear did not resume the perception of sound, then there is not sensorineural hearing loss. Any time the Rinne test results in bone conduction being better than air conduction, there is a **conduction problem**.

Selected Conduction Hearing Defects

Cerumen impaction is a very common cause of conduction hearing loss, and it is simple to understand. If there is excess earwax (cerumen) produced by the auditory canal, it accumulates. The earwax forms a physical barrier that prevents sound waves from reaching the tympanic membrane. The sound waves, intended to be directed through the air to the solid tympanic membrane, meet a rigid ball of earwax instead. The solid earwax bounces the sound waves back out of the auricle, and so the tympanic membrane never gets a chance to see those waves. The sensation is **decreased hearing** and a feeling of **fullness** in the affected ear. But because it's just normal earwax, there will be no fever or pain (distinguishing it from infectious etiologies that cause similar symptoms). Removal of the cerumen with very small tweezers and very steady hands removes the obstruction.

Otitis media is another common cause of conductive hearing loss. It was discussed in Microbiology. A bacterial infection will present with fluid in the middle ear, within the tympanic cavity, exerting pressure

on the tympanic membrane, resulting in the tympanic membrane being stretched taut and having no room in the tympanic cavity to vibrate into. Because of that pressure, the tympanic membrane cannot generate the amplification effect, as it requires the air space the fluid is occupying to vibrate. Otitis media is diagnosed by looking into the ear while insufflating a small amount of air. When the tympanic membrane doesn't budge despite the puff of air, the diagnosis is made. Other clues are the loss of light reflex and evidence of fluid, but the best diagnostic test is the insufflation of air.

Cholesteatoma describes a benign growth of oropharyngeal mucosa in the middle ear. The middle ear is normally lined with respiratory epithelium, like all extensions of the gut tube. Similarly, because it is derived from endoderm, the other epithelium the cells of the middle ear know how to make is nonkeratinized stratified squamous epithelium. Because there isn't much space in the tympanic cavity, over time, the growth erodes the ossicles, mastoid, and external auditory canal. This condition isn't common, but it does represent a tumor growing into and eroding the bones that amplify sound.

Otosclerosis is a condition of abnormal bone deposition in the middle ear, but most importantly at the **rim of the oval window** (vestibular window), which is normally a thin fibrous membrane that is required for the transmission of sound to the cochlea. The mechanism is uncertain, and the progression to marked hearing loss begins in the first two decades of life, but the loss of hearing may not occur for several more decades.

Osteopetrosis (congenital) and **Paget's disease of bone** (very late in life) were discussed in the Endocrine module. Overgrowth of bone may either close off the middle ear's connection to the cochlea (as in otosclerosis, a conduction defect) or may impinge the auditory nerve (a sensorineural defect).

Selected Sensorineural Hearing Loss

Presbycusis defines a normal age-related loss of hearing. It affects **high-frequency** hearing first, secondary to the destruction of the hair cells at the cochlear base. The patient may complain of not being able to **hear in crowded spaces** and **tinnitus** (ringing of the ears). The process is gradual and can claim more of the basilar membrane over time, starting with high frequencies and progressing to low frequencies. This is what **hearing aids** are for—amplification of the natural sound waves to create a greater vibration to accommodate for the lost signal. The pathophysiology is not elucidated and has been based less on the mechanism of illness and cellular changes, with focus mainly on audiology testing. It is good to categorize that way clinically, as audiologists—without the aid of physicians—can make diagnoses and initiate treatment. It limits our comprehension of the disease process but gets elderly patients who cannot hear, hearing again.

Noise-induced hearing loss is not the same process but can exacerbate presbycusis, as very loud noise contributes to injury and loss of the hair cells in the base of the basilar membrane. Again, the mechanism is not well elucidated, but it involves the loss of the basilar membrane tuned for high frequencies. The symptoms of noise-induced hearing loss are the same as those of presbycusis. **Ear protection** should be worn in occupations that have loud noise (construction) and in settings of purposeful volume increases (concerts, clubs). Although this isn't a proven mechanism, it makes logical sense that the basilar membrane at the entrance and exit of the perilymph-filled ducts—those nearest the stapes—would succumb first to loud noise.

Ototoxic drugs can lead to temporary or even permanent hearing loss. Administering too much of a loop diuretic at once is nephrotoxic and ototoxic. Other classic causes of both ototoxicity and nephrotoxicity are the **aminoglycosides** (antibiotics) and **cisplatin** (chemotherapy).

Acoustic neuroma, the misnomer for schwannoma, was discussed in intracranial cancers. When you see bilateral schwannomas, think neurofibromatosis type 2. No need to go deeper here.

Dizziness

Dizziness is a catch-all term used by patients to describe the experience they are having. It is an important symptom, but one that needs to be better classified. A patient's sensation of dizziness needs to be further clarified. Never write "dizziness" as a symptom, but rather elucidate what exactly the patient means—report presyncope (passing out), vertigo (room spinning), or ataxia (unsteady).

Separating **presyncope** from **vertigo** is the first key distinction. After that, **vertigo** must be distinguished from **ataxia**. Asking patients questions such as, "*is the room spinning?*" or "*are you unsteady on your feet?*" assesses for vertigo and gives a more specific sensation description. Questions like, "*do you feel like you are about to pass out?*" and "*does your vision get dark?*" achieves the same effect for presyncope. By describing the symptom concretely in the question, allowing for yes/no answers, patients are more readily able to choose the symptom, rather than describe it themselves. Ataxia is evaluated by monitoring gait and cerebellar testing (**dysdiadochokinesia**, heel-to-shin maneuvers). This section is obligatory because dizziness is so often reported by patients, but never conveys diagnostic utility. And this is dipping into very clinically oriented content. As best as we can, we will stick to cellular mechanisms of vestibular function and dysfunction.

For the remainder of this lesson, we're going to discuss **balance, vertigo, and nystagmus**, and the **vestibular portion** of the vestibulocochlear apparatus.

Vestibular Apparatus

Balance is determined by the vestibular apparatus. The vestibular apparatus is made up of the **utricle**, **saccule**, and the **three semicircular canals**. You should mentally separate the semicircular canals as sensing **rotation**, and the utricle and saccule as sensing **(de)acceleration** (lateral movement and posterior-anterior movement). Both use a **hair cell** encased in a **gelatinous mass** and send axons through the vestibular fascicle of the vestibulocochlear nerve. We handle the semicircular canals separately from the vestibule (utricle and saccule).

The semicircular ducts/canals are arranged at different angles from one another, each oriented in an x, y, z plane, the mirror image of the contralateral vestibular apparatus. The semicircular ducts are located within the canals and are filled with **endolymph**. The **hair cells** are found at the outpouching of these ducts, in the **ampulla** associated with each of the three semicircular canals. The hair cells are attached to a structure called the cupula. As the head **rotates** in space, the endolymph shifts with the movement, pushing the cupula one way or the other. This causes all hair cells attached to the cupula to have their stereocilia displaced. Some cells are displaced in the activation direction, others in an inactivation direction. It becomes activating or deactivating based on the relative opening and closure of potassium channels. The cochlea and vestibule share the same endolymph, produced by a common structure between them. Thus, hair cells are activated when the displacement of the stereocilia opens potassium channels, **depolarizing** the cell in a graded response, **increasing that cell's rate of firing**. Some cells have their stereocilia displaced in an inhibitory direction, the movement closing more potassium channels, leading to **hyperpolarization** and a **decreased rate of firing**.

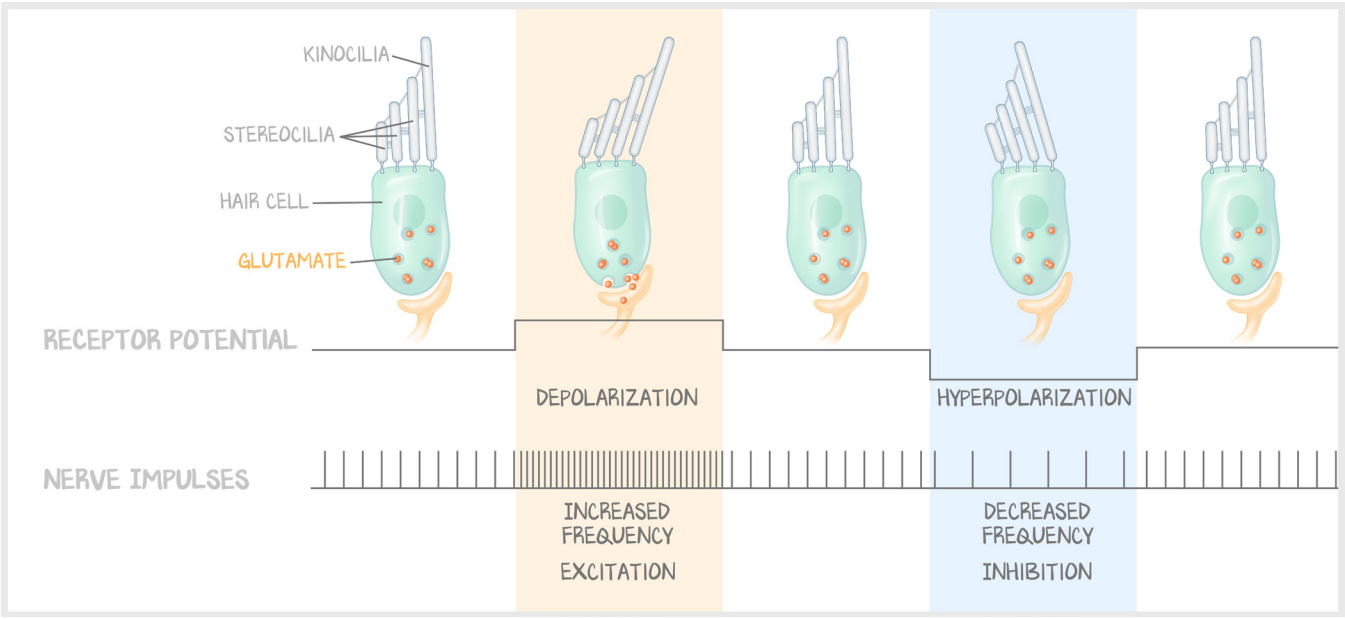

Figure 4.6: Hair Cells in Gel
Just as with the hair cells with their stereocilia trapped in the tectorial membrane, the hair cells of balance and direction can take on angles that are more obtuse (opening potassium channels and leading to depolarization) or more acute (closing potassium channels and leading to hyperpolarization). Depolarization leads to increased frequency of discharge. Unlike those in the cochlea, the cell bodies are fixed, and the stereocilia are displaced by the gel. The kinocilia are the largest, longest stereocilia.

Each cell has a graded change based on its location in the cupula. By having so many cells in a line, and the alteration of one cell slightly different than that of the cell next to it, there can be a continuous and gradual change in signal across the cupula. The sum of all the graded signals from all the hair cells of one cupula translates to sending that signal back to the brain (mainly the cerebellum). Unlike the hair cells of the cochlea, each hair cell of the vestibular apparatus has a **kinocilium**, a **leading stereocilium** that defines its polarity. The endolymph moving within the ampulla has side-to-side movement only. Hair cells are oriented with their kinocilia towards the middle of the cupula and mirror the opposite side. When the cupula moves, it will "open" one side (the kinocilia will have a more obtuse angle than at rest) just as much as it will "close" (the kinocilia will have a more acute angle than at rest) the other side. By having a continuum of cells, each cell responding in a slightly different way based on its proximity to the midline, the combined signal informs the brain of rotational muscles in one plane. When all three semicircular canals act in concert, each one providing a continuum of information for the plane it is sensing, summed together, there is an accurate depiction of rotation in all three planes.

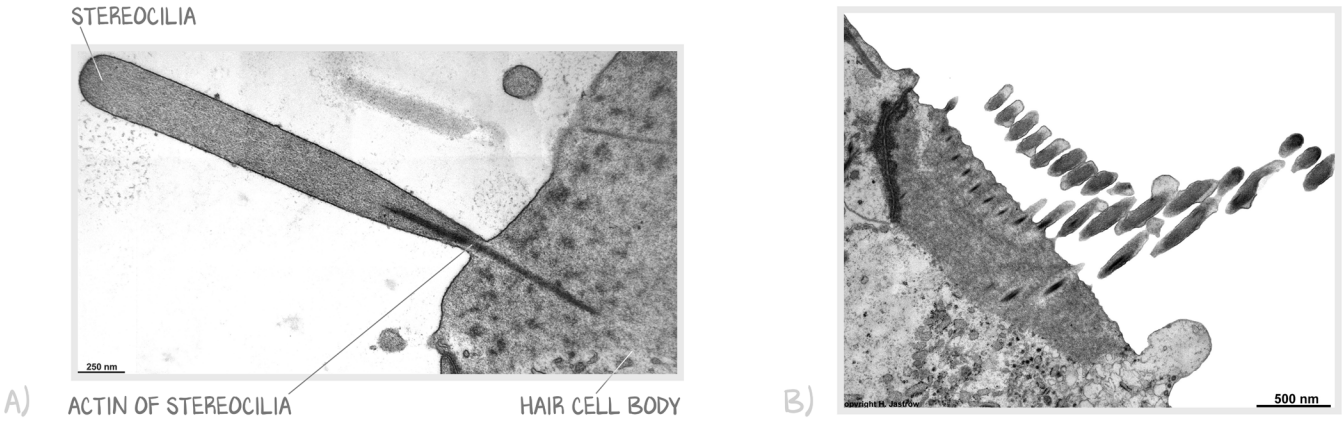

Figure 4.7: Stereocilia and Kinocilia
(a) Very-high-powered electron micrograph demonstrating a single stereocilium with its actin connection to the cytoplasm of a hair cell. (b) Lower-power electron micrograph demonstrating the orientation of the kinocilia (longest, right) with a decreasing gradation in height towards the left. These stereocilia appear to be discontinuous, but that is an artifact of the preparation and imaging—they are connected to each other and continuous from their distal tips (which are visible) to the cytoplasm of the cell below (not visible).

The opening of the kinocilia opens **potassium channels,** just like in the cochlea.

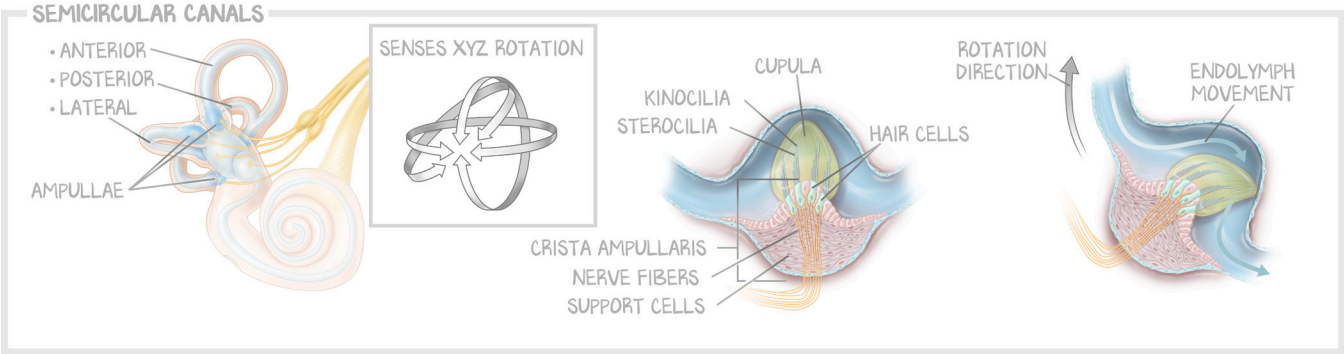

Figure 4.8: The Vestibular System—Semicircular Canals and Hair Cells in Gel
The semicircular canals, one in each of the *x*, *y*, and *z* planes, contain endolymph. In the ampulla—the base—of each semicircular canal, there is a cupula. The cupula is the gel. As endolymph moves due to rotational motion, the endolymph displaces the cupula and, therefore, the hair cells that generate the electrical signal due to increased or decreased rates of discharge. The bipolar cells connect to the neurons that become the axons of the vestibular nerve. The combination of movement of the three semicircular canals conveys rotational motion in all three planes.

The **saccule** and **utricle** determine the **acceleration** of the head in both the **vertical** and **horizontal** planes. They are yet another take on the hair cell. The saccule and utricle have a modified cupula. The hair cells have kinocilia oriented towards the medial portion of the cupula, and they are embedded in a gelatinous layer, just like in the semicircular canals. But this system utilizes physiological **otoliths**, stones made of **calcium carbonate** that are atop the gelatinous layer. These have weight, which means they can be moved with **gravity** or **acceleration** and **deceleration**. As the person changes position (seated to standing, lying down from standing) or moves in space, the endolymph allows the stone, floating in the endolymph, to be displaced by whatever forces are acting on it (gravity, locomotion from the legs). The **stones drag the gel**, dragging the hair cells. Like the hair cells of the semicircular canals, there is a continuous representation of directionality across the ampulla, the saccule taking care of the horizontal plane, and the utricle taking care of the vertical plane. Those that depolarize increase their rate of fire; those that hyperpolarize decrease their rate of fire.

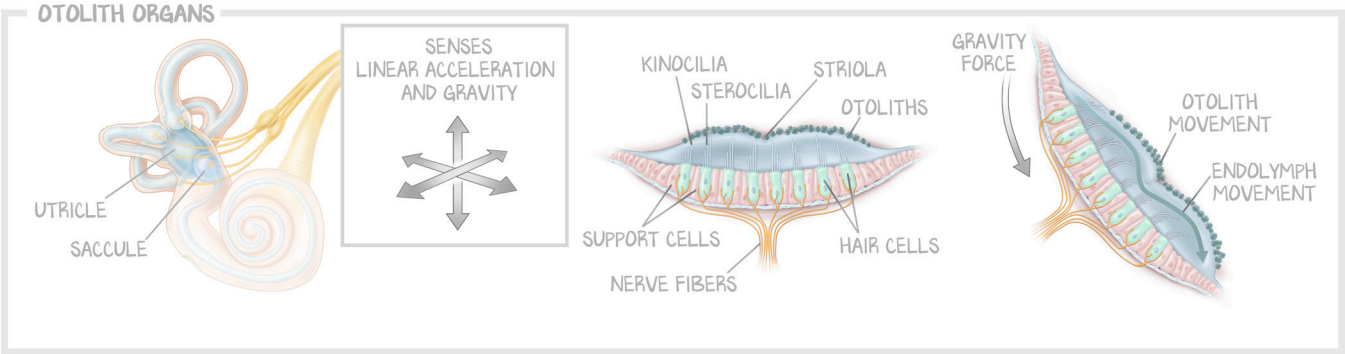

Figure 4.9: The Vestibular System—Otolith Organs and Hair Cells in Gel
Just as the stereocilia of the hair cells of the semicircular canals are displaced by rotational movement, those of the saccule and utricle are displaced by gel with rocks—otoliths made of calcium carbonate crystals—on top. The rocks on top respond to positional changes and gravity, dragging the gel and, therefore, the stereocilia of the hair cells.

Vertigo

Vertigo can be subdivided into peripheral and central.

Peripheral vertigo is caused by a defect of the vestibulocochlear nerve or the vestibular portion of the vestibulocochlear apparatus (i.e., the peripheral nervous system). Because there is a lesion outside the CNS, there will be **abnormal hearing** or **tinnitus** and **horizontal nystagmus**, but **no other focal neurologic deficits**. The most common cause of peripheral vertigo is benign paroxysmal positional vertigo (BPPV). Other causes include Ménière's, labyrinthitis, and medication side effects.

Benign paroxysmal positional vertigo is caused by sudden-onset, recurrent, and usually brief (< 1 minute) vertigo symptoms. These are usually provoked by positional changes in the head. They are caused by a displaced **otolith**. Otoliths are used by the saccule and the utricle. **Endolymph is continuous** between the semicircular canals, utricle, saccule, and cochlea—all endolymph comes from the same place. So if one of those calcium carbonate stones gets displaced from the cupula of the saccule or utricle, that stone could make its way into the semicircular canals. There, the stones still have weight, and so can manipulate the stone-free cupula of the semicircular canals. When the person moves, the stone adds a directional signal that shouldn't be there. The vestibulocochlear apparatus on one side sends incongruent information because of the stone, different from that of the unaffected side. The brain puts these two signals together, and the cortex isn't sure which one is right. So, sensing both, the patient suffers vertigo. Much like the inability to appropriately focus light in refractory disorders of the eye, the incoming inputs produce a "blurry picture" of where the head is, and whether it is rotating. The **Dix-Hallpike maneuver**—the examiner, dropping the patient from a 45-degree angle to flat (stretcher maneuver, which can also be done with hands and not a stretcher), then turning the head—is positive if there is **rotary nystagmus**. Rotary nystagmus is the result of the misinformation from the otolith. The pupils will rotate, and the patient gets vertigo. The treatment is the **Epley maneuver**, successive manipulation of the patient and leaving them to rest, working the stone out of the semicircular canals, and back into the vestibule where it belongs.

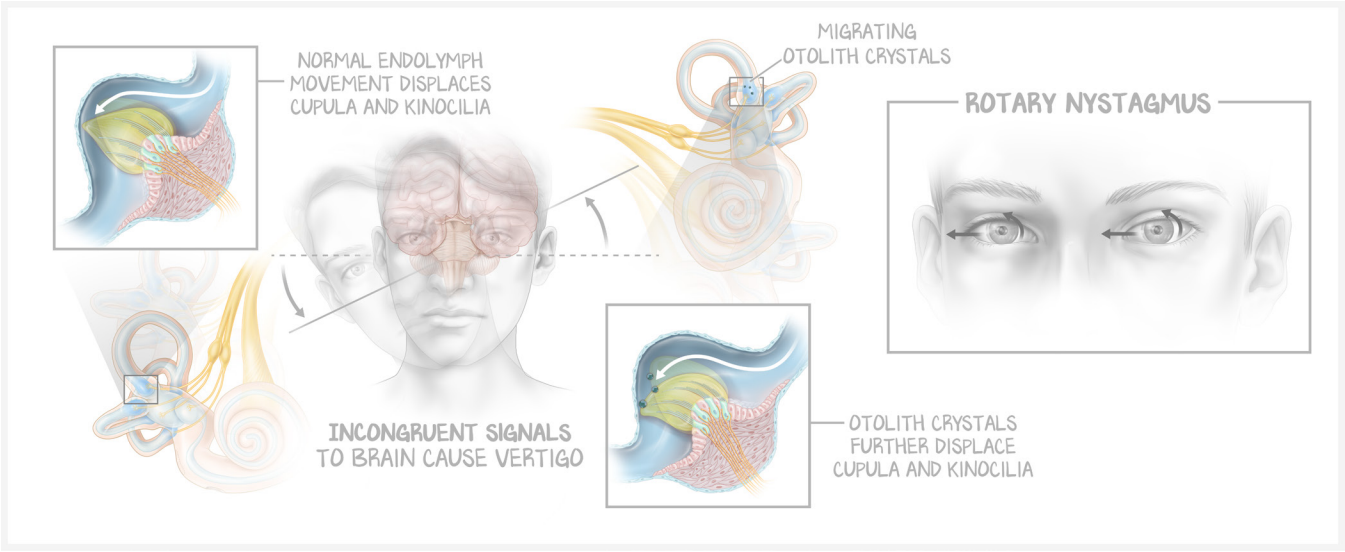

Figure 4.10: Otoliths Can Cause BPPV
When an otolith inappropriately makes its way out of the gel of the saccule or utricle and gets into the gel of a cupula, the added weight—the added effect of the otolith—causes vertigo. It isn't just the presence of the otolith—if there were only one vestibulocochlear apparatus, there would be no mixed signal. But because the brain needs inputs from *two* vestibulocochlear apparatuses, if one communicates information correctly while the other doesn't, the incongruent signal causes the perception of vertigo.

Ménière's disease classically presents with the triad of **vertigo**, **tinnitus**, and **hearing loss**. It is caused by excessive **endolymph pressure**. Like much of what we've seen in Neuroscience (specifically CSF and orbital aqueous humor), the cause of excess pressure tends to be increased resistance to outflow. Unfortunately, there isn't a simple solution (like iridectomy for glaucoma) or even a complicated solution (like a VP shunt for CSF) for Ménière's. The goal is to keep total body sodium reduced, with the use of **low-salt diets** and **gentle diuretics**. We'll cover labyrinthitis and vestibular neuritis in Clinical.

For all causes of peripheral vertigo except Ménière's disease, the treatment is symptomatic rather than treating the underlying cause. **Vestibular suppressants** (which we saw in the Gastrointestinal module), such as antihistamines (**meclizine**), benzodiazepines, and antiemetics, can be used in conjunction with other forms of therapy for temporary symptomatic relief. Vestibular and balance rehabilitation therapy (VBRT) is also effective in the treatment of various forms of dizziness. It is often performed by physical and occupational therapists.

Central vertigo is caused by the loss of neurons or tracts—**stroke** being the most common cause, **tumors** being another—of any structure involved in balance—the vestibular nuclei, thalamus, cerebellum, etc. The causes of central vertigo are usually more catastrophic, and so will present with **other neurologic defects** or **seizures**. Central vertigo necessitates brain imaging. Peripheral vertigo doesn't guarantee imaging isn't involved, but most of the time (except schwannoma), brain imaging isn't required. In central vertigo, **any nystagmus is possible**, but often at this stage of training, the dogma is: "*rotary, BPPV; horizontal, peripheral vertigo; vertical, central vertigo.*"

Citations